Blindness in an American Boy Caused by Unrecognized Vitamin A Deficiency

Vitamin A deficiency is an enormous problem in the developing world, where UNICEF and other organizations now distribute more than 400 million high-dose, biannual supplements every year. In developed countries vitamin A deficiency is rarely found. Xerophthalmia caused by vitamin deficiency has been reported in food faddists and psychiatric patients. However, most of the cases observed in developed countries are due to alcoholism and conditions causing malabsorption. Since better nutritional standards in affluent societies make vitamin A deficiency rare, early diagnosis of such deficiency may be overlooked. We report a case of bilateral keratomalacia caused by vitamin A deficiency, leading to bilateral irreversible blindness in a 6-year-old Asian boy living in New York, NY.

Report of a Case. A 6-year-old Asian boy was referred to the Cornea Service of the Massachusetts Eye and Ear Infirmary, Boston, in December 2002 with a 10-month history of bilateral corneal ulceration. The patient was sent to the Retina Service for consultation. Ultrasonography showed a total cicatricial retinal detachment in the left eye, while only a traction detachment in the right eye that, to one examiner, appeared to have “swollen” eyes and gradual vision loss after a classmate “splashed chocolate” in his face. He was brought to the emergency department of that institution, where he was given antihistamine eyedrops, and was discharged home with a diagnosis of an allergic reaction.

The parents subsequently took the child to a Chinese herbalist who gave the patient unknown eyedrops for the next 2 days, with improvement of the child’s redness and swelling. The parents thought no further medical advice was necessary. Three weeks afterward, however, they noticed that the boy did not want to get out of bed because of photophobia and decreased vision. The parents brought him to another medical institution in March 2002, where the physicians discovered that the patient was on a strict vegetarian diet also followed by the parents (no meat, no eggs, no dairy, and no fish). The boy had narrowed his diet to potato chips, rice, soy milk, and tofu.

On examination, the child was not able to identify objects, and ophthalmologic examination under anesthesia showed bilateral corneal perforation and conjunctival keratinization with aqueous leaking from the left eye. The patient was taken to the operating room for bilateral debridement of the corneal ulcers and corneal transplants. The postoperative note reported total corneal melts, epithelial ingrowth, extrusion of vitreous, and choroidal and retinal detachments in both eyes. The patient received 0.3% tobramycin and 0.1% dexamethasone sodium phosphate postoperatively.

The patient was found to be severely vitamin A and zinc deficient, as well as mildly protein deficient. Vitamin A level was undetectable, zinc level was 45 µg/dL (6.9 µmol/L) (reference range, 57-113 µg/dL [8.7-17.3 µmol/L]), and vitamin D level was 3 pg/mL (7.8 pmol/L) (reference range, 8.9-47.0 pg/mL [23.1-122.2 pmol/L]). He received 5 days of high-dose vitamin A supplementation and 12 days of zinc supplementation, as well as high doses of cholecalciferol. The boy also had macrocytic anemia on admission, and therefore cyanocobalamin and iron supplementation was started. He also received treatment for thrombophlebitis of his right foot. He was discharged after 3 weeks of hospitalization.

On ophthalmic evaluation at Massachusetts Eye and Ear Infirmary in December 2002, the patient had a visual acuity of no light perception in both eyes, extraocular motility was full in both eyes, and the eyes were soft by palpation. Slit-lamp examination showed areas of corneal thinning and ectasia, severely vascularized and conjunctivalized cornneas, and no view of the anterior chamber. The patient was sent to the Retina Service for consultation. Ultrasonography showed a total cicatricial retinal detachment in the left eye, but only a traction detachment in the right eye that, to one examiner, appeared to have some light perception (Figure 1).

The patient came for follow-up 6 months later, and light perception in the right eye was confirmed. B-scan ultrasonography was identical to that from his previous visit. The patient underwent an attempt at surgical rehabilitation of the right eye including removal of the conjunctival flap, penetrating keratoplasty (8.5 mm/8.0 mm), intracapsular cataract extraction, and anterior vitrectomy. One month after surgery, visual acuity remained light perception in...
the right eye and no light perception in the left eye. Slitlamp examination showed an edematous graft in the right eye and an opacified, vascularized cornea in the left eye. Renewed B-scan ultrasonography in the right eye showed that the patient now had total retinal detachment in a funnel configuration. The left eye showed a tight funnel retina as previously seen. This configuration was considered inoperable, and given the extent of the patient’s anterior segment problems, further surgery was deferred.

Comment. Xerophthalmia (xerosis, dry; ophthalmia, inflamed eye) is a term that includes all ocular manifestations of vitamin A deficiency (night blindness to keratomalacia) and has been categorized by the World Health Organization.

The differential diagnosis of keratomalacia includes severe sicca syndrome, exposure keratopathy, or corneal ulcer (infectious, neuropathic, or autoimmune). Xerophthalmic ulceration ranges from small, characteristically sharp-margined ulcers located in the periphery of the cornea, to full-thickness, nearly limbus-to-limbus melting. The mechanism of corneal necrosis remains unclear, but it has been postulated that inflammatory cells releasing proteases such as collagenases may be responsible for the corneal necrosis.

When keratomalacia has progressed to almost total melt involving the entire cornea, vitamin A treatment has virtually no effect. Herein we have described the unexpected tragic evolution of a case of vitamin A deficiency in a boy from a highly educated, affluent family that was diagnosed late and led to blindness despite the best medical effort. This case reminds us once again that social customs, cultural differences, and lifestyle matter in making an accurate and prompt diagnosis.

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The authors have no relevant financial interest in this article.

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Capsular Bag Hematoma Following Trabeculectomy

Since its introduction in 1967, trabeculectomy has become the standard surgical treatment modality for most forms of glaucoma. The early postoperative complications reported are hypotony, shallow or flat anterior chamber, hyphema, choroidal detachment, uveal effusion, wound leak, malignant glaucoma, suprachoroidal hemorrhage, and endophthalmitis. We describe an interesting case of a capsular bag hematoma following trabeculectomy, a hitherto unreported complication.

Report of a Case. A 57-year-old man was initially examined at our tertiary care institute and had a history of total visual loss in his left eye following surgery for glaucoma elsewhere 3 weeks earlier. On examination, his best-corrected visual acuity was 20/30 in the right eye, and hand motions close to his face with accurate projection of rays in the left eye. Intraocular pressure in the right and left eyes was 14 and 12 mm Hg, respectively. The left eye revealed a thin moderate-sized bleb and a quiet anterior chamber with normal depth. Results of a dilated examination revealed a posterior synchia at the pupillary margin at the 1 o’clock position and a peripheral iridectomy in the same meridian. The crystalline lens appeared to have a brownish hue throughout, with a bright red collection in the anterior subcapsular area just behind the area of posterior synchia (Figure, arrow). There was no view of the posterior segment. Findings from a B-scan ultrasonogram revealed a normal posterior segment in the left eye. The patient was posted for phacoemulsification and aspiration of the blood along with intraocular lens implantation. Capsular staining with trypan blue failed to provide adequate contrast in view of the dark reflex of intralenticular contents. Capsulorhexis was then achieved from the anterior capsule reflex under high magnification. Phacoemulsification power was totally ineffective in removing the blood-impregnated epinucleus shell, and it had to be manually stuffed into the port of the phaco tip with a chopper. A normal red reflex was achieved as soon as this blood clot was removed, and